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ON SCLERODERMA AND CHRONIC RHEUMATOID ARTHRITIS.[1]

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Our knowledge of some of the trophic diseases is as yet so obscure that new observations concerning them cannot but be of value. Several of them, especially acromegaly and myxœdema, have in the past few years received a large share of attention. Scleroderma, on the other hand, has not been studied to the extent that its importance deserves. This is also true of that other obscure disease, chronic rheumatoid arthritis. Repeated observations have convinced me that as regards scleroderma, our conception is as yet too limited. Our point of view has in the past undergone several important changes. The first was the recognition that all forms of local scleroderma and general or diffuse scleroderma are one and the same affection. The second consisted in the recognition of the fact that the disease process is not necessarily limited to the skin, but may include other structures as well. In a paper,[2]

[1] Read at the twenty-fourth annual meeting of the American Neurological Association, May, 1898.

[2] Journal of Nervous and Mental Disease, July, 1896.

read some two years ago before the Neurological Section of the New York Academy of Medicine, I laid emphasis upon the fact that tendons, muscles, fasciæ, bones and joints may also become involved. That this involvement of tissues other than the skin may, in rare instances, be excessive, there can be no doubt. Indeed, the question arises whether they may not at times exceed those in the skin. The changes in the latter have so long been regarded as the primary and all-important features of the disease, that changes in other structures have not only been looked upon as secondary, but even as dependent upon the changes in the skin. Thus, most writers refer the restricted movements of joints or the atrophy of bone, as seen in the fingers in sclerodactyle, to the contracture of the overlying dermal structures and to the interference with nutrition thus produced. For instance, the impairment of movement in the fingers, of the wrist and of the elbows, appears in many cases to be directly dependent upon the contraction of the skin. In sclerodactyle, again, the fingers become for the most part thin and tapering, and when such cases are skiagraphed, we may find that the phalanges, especially the distal phalanges, have become decidedly pointed or sharpened, as though they were undergoing atrophy.[3] In such cases the atrophy of the bone appears to be general, and it does not seem unreasonable to suppose that the changes in the bones are directly dependent upon the changes in the skin. The latter may be dense, hard and contracted, and may interfere greatly with the blood supply of adjacent structures. We should bear in mind, however, that changes may occur in phalanges when the skin is not hard and contracted, but merely thin and atrophic. This was true of a case studied by me several years ago, and which I have already reported.[4] In this case the proximal and middle phalanges

[3] Loc. cit. Addendum.

[4] Loc. cit.

of the little finger of the left hand had almost entirely disappeared, while the skin was much wasted but not infiltrated, the finger being abnormally mobile because of the loss of bone. Many of the phalangeal joints also were ankylosed, the ankylosis being evidently due to fixation of the joints independently of the changes in the atrophic skin. Again, that changes occur in the bones of the fingers, independent of contracture and atrophy of the skin, is proven beyond all doubt by the following case.

Case I.—A. G.; female; married; aged 44 years; housekeeper.

Family history, negative; father, mother, several brothers and sisters all living and well; no history of nervous or skin affections.

Personal History.—Was perfectly well as a child and young girl; lived out as a servant; was compelled to do a great deal of washing, and frequently worked in the open air with her hands and face exposed, while wet, to intense cold; menstruated at fourteen; married at thirty years, but never became pregnant.

When twenty-six years of age, she noticed that the middle and ring fingers of the right hand were becoming stiff and swollen, and that this condition was more marked at times. A little later the affection made its appearance in the corresponding fingers of the opposite hand, and, finally, all the fingers and both thumbs became involved. Very soon the stiffness and swelling became decided, and the fingers became chronically swollen and thickened, so that it was impossible to flex or extend them in the normal manner. About this time also the face became hardened and stiffened. Distinct swelling does not appear to have been present. There appeared to be merely an infiltration, followed by a gradual shrinking of the skin. The lips became much thinner, while the skin of the cheeks and forehead was tense. Gradually, also, infiltration of the skin of the back of the feet, and slightly of the toes, became noticeable. This, however, has never been as marked as in the hands.

At various times small trophic ulcers made their appearance upon the first phalangeal joint of the little finger, and, after having existed for some time, slowly healed. Gradually the finger ends became more swollen than the rest of the finger, so that the latter became club-shaped. At the same time the tips of the fingers became shorter. Ulcerations

frequently occurred about the root of the nails. The nails became shortened and flattened. Upon one digit, the right forefinger, the nail was lost altogether. These ulcerations, the patient states, were always very painful.

Some time ago the teeth of the upper jaw gradually loosened, one by one, and had to be removed.

In other respects her health has been fairly good. She has suffered at times from mental depression, and quite frequently from headache. Vertigo and tinnitus were not at any time present.

Menstruation, which had been regular and normal up to

FIG. I. Sausage-shaped fingers in sclerodactyle with trophic changes in nails and distal phalanges.

two years ago, had ceased at that time, and had not since returned.

Status Præsens.—Patient presents the facies of scleroderma. The skin of the cheeks, forehead and lips is tensely drawn. The forehead presents an erythematous flush. Persistent pressure produces marked pitting over the forehead. No pitting can be elicited over the rest of the face. When the patient talks or smiles, it is readily seen that the mouth is much contracted; the lips become tense and thin. There is decided palor of the tongue, roof of the mouth and fauces.

There are no changes in the trunk or limbs other than

those described, save in the skin over both shoulders, which is somewhat infiltrated and hard. There is everywhere an absence or diminution of the superficial fat. In the hands there is slight infiltration of the dorsum and very marked infiltration of the fingers and thumbs. The fingers are more or less fixed in the semi-flexed position, and are thick, sausage-shaped or club-shaped. The distal phalanges are evidently shortened by atrophy. All of the fingers reveal traces of pre-

FIG. II. Skiagraph of the fingers of the right hand showing changes in the distal phalanges.

vious ulceration in the matrix of the nail, save the little fingers, the nails of which are apparently normal. On the forefinger of the right hand the nail has been entirely lost, a scar of the matrix only being left. An ulceration is at present active at the root of the nail of the left thumb. This ulcer is exceedingly painful. Tactile, thermal and pain senses are everywhere preserved. Knee-jerks are not changed. An examina-

tion of the blood failed to reveal any evidences of leucocytosis. Examination of the urine, negative.

Some two months after the first examination the patient again presented herself with an ulcer over the left olecranon, while extreme ulceration had recurred on the fingers. Ulcers were present on all of the fingers, the little fingers being this time also affected. The ulcers involved the matrix of the nails and also the tips of the fingers. They were all very painful.

Seen again four months later, the ulcers were evidently in process of healing. The little fingers had become much contracted, and the mouth was also more drawn. Backache was also complained of, and the general health had evidently become much impaired. The patient complained also of cold sensations and occasional flushes.

FIG. III. Skiagraph of the left thumb and forefinger.

In this case the changes in the face were so typical that there could be no doubt as to the nature of the affection. The face was drawn, the cheeks flattened, the angles of the mouth slightly drooping and the lips contracted. The hands were also in a condition of sclerodactyle. All of the digits were fixed and rigid, but instead of being pointed and showing excessive contracture and atrophy of the skin, they were, as is seen in the photograph (Fig. I.), enlarged, bulbous and sausage-shaped. Ulceration, as has already been described, had taken place in the tips of the digits and thumbs, with loss of the soft tissue and also with loss of some of the nails. When these fingers were skiagraphed (Figs. II. and III.), a most interesting condition of the bones was revealed. The changes were limited to the distal phalanges. There was not the general sharpening and wasting shown in the first case skiagraphed

by me[5], but, instead, there had ensued a gross and very decided loss of bony tissue, and in several of the digits, for example the thumbs, in which the nails had been fairly well preserved, and in which there had been no wasting of the pulp of the tip, very striking changes were revealed in the bones. The changes were of such a character as to justify no other inference than that they were trophic in character. In this connection it is interesting to recall the fact that Wolters described, in a case of sclerodactyle, which he examined microscopically, an interstitial inflammation of the phalangeal bones. As is well known, symptoms suggesting Raynaud's disease are every now and then observed in selerodaetyle. In this case, however, no such symptoms were present, and there was no history suggesting vascular crises. The case is further interesting because the trophic changes in the fingers strongly call to mind those of Morvan's disease. From the latter affection, however, it is sharply defined by the absence of all sensory losses, all forms of cutaneous sensibility being preserved, and by the presence of pain in the ulcers.

The following case which must be placed under the caption of rheumatoid arthritis—whatever that may be—presents a number of features pointing strongly to scleroderma, and suggesting a similarity in the pathological changes at work. The case is specially interesting when we reflect that it is not improbable that under chronic rheumatoid arthritis, two or more clinical entities may be confused.

Case II.—E. McG., male; aged 28; born in this country; inmate of nervous wards, Philadelphia Hospital.

Family History.—Father died of pneumonia at 46; mother living and well; one brother and three sisters living and well; one brother and two sisters died in infaney; no history of rheumatic, skin or nervous affections in the family.

Previous History.—Was well during childhood, save that he frequently had attacks of croup; also had measles; frequently suffered from headache; had good health otherwise up to fifteen years. At that time had a swelling of the right knee, which confined him to bed for one week; the attack was not accompanied by pain. It was pronounced, according to patient's statement, to be "rheumatism and white swelling." One year later the swelling recurred, and he was confined to

[5] Loc. cit. Addendum.

bed two weeks. One and a half years later the attack again recurred; this time it lasted three months, and was accompanied by pain. Seven months later, in June, 1887, he suffered from another attack. The right ankle began to swell, and became painful, so that he could not walk. This time the attack lasted three weeks, but subsequently he became, as he thought, entirely well. Some time later, however, he began to feel stiff all over; had occasional "catching" of the muscles of the left thigh, half way between hip and knee. He would be so stiff at times that he could not walk. His general strength also suffered severely; weakness became marked. In about a year—1888—he was able to use his legs but little on account of stiffness, though this stiffness varied considerably from day to day. Both feet also became somewhat swollen

FIG. IV. (Case 2.) Showing general position of limbs.

in the daytime, the swelling disappearing on going to bed. His arms, neck and back also began to get stiff about this time, and these symptoms gradually grew worse. He was treated at various times with massage, but passive movements gave him pain. For a time, in 1894, his hands were swollen, but this swelling subsequently disappeared. About this time, also, he suffered severely from neuralgic pains in the head. Little by little the stiffness increased, until his limbs, hands and feet became more or less fixed in position. About three years ago the skin of the legs became smooth and shiny, and subsequently the skin in other situations assumed a similar character. About this time, also, the jaw became quite stiff. He has not been able to feed himself for five years. He has been unable to walk or even stand up for eight years.

Status Præsens.—Patient lies extended upon his back in bed. Some diffuse muscular wasting, with loss of superficial fat, has evidently taken place. No local muscular atrophies are, however, apparent. Both legs are in a position of extension, the feet in extreme extension. The amount of voluntary movement in the legs is extremely limited. The patient is able, by the action of the thigh muscles, to move legs upward to an extremely limited degree—a mere fraction of an inch. On attempting to make passive movements with either leg, it is found that the ankle joint, tarsal and metatarsal and

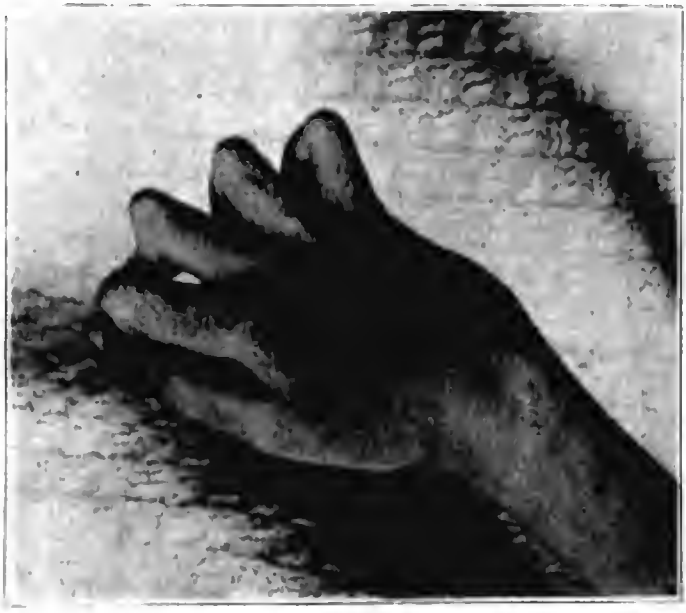

FIG. V. Right hand in Case 2.

knee joints are absolutely fixed. There is also fixation of both hip joints, though here the fixation is not absolute. Very slight movement in antero-posterior direction, as just stated, can still be made. On attempting lateral movement, however, the limbs cannot be moved without moving the entire pelvis. The right knee joint is decidedly enlarged, the patella quite prominent. The circumference of the right knee joint is 12⅝ inches, of the left knee joint 11⅛ inches. The right leg is not so completely extended as the left; it is very slightly flexed at the knee. The right foot is fixed in extension; its toes are in a position of marked abduction and slight flexion. The right

tarsus is thickened. The left foot is also fixed in extension; its great toe is extremely flexed, but not abducted or adducted; its second toe is markedly adducted and crossed over the great toe; the remaining toes of the left foot are slightly adducted, and neither flexed nor extended.

The arms are extended, but not completely so. There is absolutely no movement at the elbows or at the wrists. There is very slight voluntary movement at the shoulders in all directions. Upon passive movement, it is found that the excursion of the arm is considerably greater than by voluntary movement, and also that there is more movement in the right shoulder than in the left. The movement does not, however, take place in the shoulder joint, or, at least, only to a very small extent, for, on attempting to ab-

FIG. VI. Left hand in Case 2.

duct or adduct the right arm, the scapula at once moves with the humerus; this is even more marked in the left shoulder. The patient is able to perform very feeble movements of flexion and extension in the fingers of the right hand. The fingers are much distorted. The proximal phalanges of the fingers of the right hand are in a position of overextension, while the middle and distal phalanges are in marked flexion; these peculiarities are most marked in the little and ring fingers; the middle and forefinger are also extended, but to a less degree. The middle phalanges are flexed at a right angle, while the distal phalanges are again in a position of extension. The fingers are spread

apart at their metacarpal articulations, the little finger being much abducted, while the forefinger is decidedly adducted, as are also the middle and distal phalanges of the fore and middle fingers. The thumb is extended, adducted and displaced, so as to occupy almost the same plane as the palm of the hand. Passive movements about equal the small voluntary movements described. In the left hand the fingers are likewise much distorted; the thumb is extended and adducted; the proximal phalanges of the ring, middle and fore finger are flexed, especially the latter; the middle and distal phalanges are also flexed in varying degrees; the middle finger is adducted so as to cross the tip of the thumb; the little finger is in the position of moderate flexion at the middle phalanx. The palmar surfaces of both hands are much indurated, and hard to the touch. Both wrists are thickened.

The head and the entire trunk are rigid in extension. The head can be slightly raised from the pillow; slight extension of the head backward is also possible. Movement of the entire head is, however, so limited that when the pillow is withdrawn the head remains fixed and extended upon the trunk. There are almost no lateral movements of head, and very slight rotary movement. The jaw is so firmly fixed that the teeth can only be separated for a quarter of an inch.

The trunk is so fixed that it is impossible to obtain any movement in any direction. The ribs are almost completely fixed in position; no movement is perceptible in any but the false ribs, and in these only upon forced respiration. The thorax everywhere feels firm and resistant to touch, the intercostal spaces offering almost as much resistance to touch as the ribs.

Respiration is almost exclusively abdominal. The abdominal muscles move freely only upon forced inspiration. They feel firm, hard and resistant to the touch, suggesting almost the board-like feel met with in tetanus. The abdominal reflexes are much exaggerated.

The muscles of the neck feel excessively firm and hard, with the exception of the right sterno-mastoid, which is soft and flaccid. The right deltoid, even in a flaccid condition, seems to have its density increased, and on efforts to move the arm it becomes excessively hard. The biceps and triceps are flaccid, yet likewise present a firm, fibrous feel. This is true also of all the muscles of the forearm. In the muscles of the left arm the same condition is noted as in the right. The right deltoid, biceps and triceps, even in the relaxed condition, feel firm and fibrous to the touch. The belly of the biceps feels as though it were made up of a bundle of coarse,

hard cords (giving a rope-like feel). The condition of the forearm is similar to the right. The muscles of the right thigh feel firm and fibrous. This is also true of the muscles of the leg, the sensation which is given to the finger being similar to that given by the arms. The condition of the left thigh and calf is practically identical with that of the right. The adductor and calf muscles, though relaxed, are, nevertheless, dense to touch, giving here and there the fibrous feel noticed in the biceps.

The face is smooth, the normal folds and wrinkles being somewhat obliterated; the surface of the forehead, nose, anterior aspect of cheek, temples and ear is smooth and glistening. The ears present a wax-like hue or gloss; indeed, they might pass as artificial models of the human ear in wax; this is especially true of the right ear. The lips are purple-pink in color. The vessels of the eyelids are much increased in number, so as to suggest a telangiectatic condition. Both cheeks are tinted a purple-pink hue, and, upon close examination, minute vessels are seen. The ears are tinted with the same hue, save in the prominent portions of the cartilage, such as the tip of the tragus and antitragus and the edges of the helix and antihelix, which are dead white.

The scalp and the neck, especially the back of the neck, reveal copious masses of epithelial scales or encrustations. These scales, the patient tells us, accumulate rapidly. They can, as a rule, be removed with difficulty both by washing and by gentle scraping, leaving a raw and slightly sensitive surface, probably the sensitive layer of epithelium, exposed. These patches are observed also back of the ears and at various parts of the chest and abdomen. The hands also present yellowish stains on palms and flexures of fingers, due to similar epithelial deposits. The backs of the hands and fingers reveal much pigmentation. The skin of the face is everywhere movable, though this mobility is restricted; this restriction is especially noticeable on the forehead. The scalp also is very restricted in movement, the loss of mobility being such as to suggest a very tightly drawn skull-cap. The fibro-cartilage of the ears seems denser, more resistant, and less flexible than normal. The cheeks are somewhat flattened. There is evident loss of superficial fat above and below the zygoma. The mobility of the facial muscles is well preserved. The skin of the eyelids, both upper and lower, seems somewhat thinner than normal. The eyelids and the sides of the nose at the root are slightly bluish in tint.

The neck and shoulders also present a glossy appearance. A number of telangiectatic patches are observed upon the

neck, shoulders, scapula and chest. The skin of the trunk is less smooth and more normal to the touch than that of the face and neck. Its mobility, however, is much diminished, and it is much firmer to the feel than normal skin. It is slightly more movable over the lower portions of the chest and abdomen than over upper portions of the chest. Tache cérébrale is noted freely over chest and abdomen.

The skin of the left thigh, especially in the middle third, appears to be slightly less mobile and more tense than normal. The middle and lower third of the thigh present the same yellowish encrustation of epithelium noted elsewhere. These

FIG. VII. Feet in case 2.

having been partially removed, the underlying skin presents numerous pink punctuate markings. The skin of the left knee, anterior aspect of the leg and foot, is also very much thinned and atrophied, and is highly glazed and shining. The skin over the foot is so thin that it permits the muscles and tendons beneath it to be plainly seen. The skin of the middle and lower third of the leg and of the dorsum of the foot presents a marked veining (the veins are apparently readily seen owing to the atrophied condition of the skin). The skin of the leg, especially in the lower portions, is much less movable than the skin of the thigh; it is especially tense over the dorsum

of the foot. The skin is especially thin over the first phalangeal articulation. The nails are much thickened, yellow, opaque and distorted; the matrix of the great toe is especially thick.

The right thigh is in every way similar to the left. Over the knee the skin is very tense, shining and thin. On the outer aspect of the leg and dorsum of the foot, the skin has the same appearance as in the left limb. The toes are glazed, and the nails are in the same condition as in the left. The atrophied changes of the skin also appear to be equally marked over the right foot, so that the tendons, veins and other structures can be readily seen through it.

The skin of the back of the neck, trunk and buttocks presents similar changes to those noted elsewhere, save that they are less marked. Its mobility seems lessened, and it seems denser to the feel than normal, but these factors are far less marked than in the skin over the anterior portion of the trunk. The soles of the feet are distinctly indurated, though less so than the palmar surfaces of the hands. There is no special loss of the plantar fat. An occasional fibrillary twitch, or twitch of a tendon, is noted in the hands. The palms of the hands present an indurated feel.

Pain is caused by attempts at passive movement or by turning the patient in bed, but this pain seems to be due to the strain placed upon the fibrous tissues and muscles, rather than to a strain upon the joints.

No sensory loss can be discovered, and, according to the patient's statement, none ever existed. The special senses are also normal. The growth of hair over the scalp, pubis and axilla and over the general surface of the body appears normal. The growth of beard is scanty. The patient swallows without difficulty liquid and semi-fluid food. No decided changes are noted in the vessels. The sound of the heart, however, is somewhat accentuated, while the pulmonary valves are distinctly roughened. No other visceral changes are observed. Bowels are constipated. Anal sphincter normal. The control over the bladder is slightly diminished; sometimes soils his bed before the urinal can be brought. The urine is normal in quantity. The solids are somewhat diminished, the urea, for example, varying from 230 to 270 grains daily. One specimen of urine revealed also the presence of peptone, though this observation was not repeated.

A blood examination by Dr. A. E. Taylor failed to reveal any evidence of leucocytic degeneration or karyokinesis.

The patient has never had sexual intercourse, has never had any seminal emissions; has occasional erections. Pubis is covered with a dense epithelial covering, similar to that noted on scalp and elsewhere.

When we analyze this case, we find that its clinical history, especially the mode of onset, accords closely with that frequently met with in chronic rheumatoid arthritis. The recurrent attacks of pain in the right knee, which appeared to be the starting point of the case, are exceedingly suggestive. Further, while trophic changes, involving the skin, nails and muscles, are not common in rheumatoid arthritis, they may occur. The case before us, however, is distinctly unusual in the extent to which these changes are present. We need only point to the tissues of the scalp, the fibro-cartilage of the ears, the tissues of the plantar, and especially of the palmar surfaces, all of which are indurated, much denser and less mobile than normal, and are evidently the seat of some diffuse sclerotic change. When we examine the muscles, the same fact is again apparent. As regards the muscles of the extremities, which are, of course, related to joints, it is conceivable that the changes found in them are arthritic in character. However, it is somewhat striking that the usual disproportion in the involvement of the flexors and extensors is not present. In purely arthritic muscular atrophy, the extensors suffer so greatly as to lead to marked flexion and contraction of the legs and arms; and, indeed, this is the usual position assumed in typical rheumatoid arthritis. Again, the diffuse induration of the muscles, very marked in certain situations, is a feature of the present case which must not be lost sight of; and even if we assume an arthritic origin for the muscular changes, how are we to explain the induration of the muscles which are not related to joints, such as the intercostals and the muscles of the abdomen? These, as pointed out, have an induration that is boardlike.

The deformity of the hands in this case also departs from that usually met with in rheumatoid arthritis. The so-called ulnar deflection is entirely absent. It is only in the toes of the right foot that abduction is seen. In the condition of the skin and nails, the hands and feet strongly

call to mind the sclerodactyle of scleroderma. Rigidity of the spine, so marked in this case, may also be met with in scleroderma, though much less pronounced. It was markedly present in a case of scleroderma described by me and reported in a former paper. In this instance the rigidity of the spine was not related to the skin of the back or of the neck, and appeared to be dependent upon involvement of the articular joints. In the same case, also, there was undoubted involvement of one shoulder joint without any involvement of the surrounding skin. Further, Legrange observed in sclerodactyle, loss of articular cartilage and calcareous deposits in the fibrous tissues, while Verneuil and Mirault observed short fibrous bands extending between the apposed joint surfaces in which there had also been destruction of the synovial membrane. While bone and joint changes do undoubtedly occur in scleroderma, and independently at times of the changes in the skin, such extensive joint changes as observed in the present instance, have not been observed in cases described as scleroderma. However, the case is extremely interesting as suggesting a general sclerotic process bringing about extensive changes in bones, joints, muscles, tendons and skin, similar in character to those seen in scleroderma. It would almost seem as though in scleroderma the process expended itself primarily upon other structures, while in this case, call it rheumatoid arthritis, if we may, the process had expended itself primarily upon the deeper structures. The study of this case suggests the further thought that under the name, chronic rheumatoid arthritis, are properly included two, if not more, clinical entities; one a disease in which a sclerotic process, similar to that which occurs in scleroderma, is active, and another in which the joint changes are attended by absorption of cartilage, eburnation of bone, osteophytic deposits, and secondary arthritic muscular atrophy.